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SUBMITTED: 2001-12-03 15:03:29
PRINTED: 2001-12-04 17:01:52
REQUEST NO.: NIH-10082248
SENT VIA: LOAN DOC
5178527

NIH Fiche to Paper Journal

TITLE:	JOURNAL OF BONE AND JOINT SURGERY. AMERICAN VOLUME
PUBLISHER/PLACE:	Journal Of Bone And Joint Surgery Boston Ma
VOLUME/ISSUE/PAGES:	1993 Feb;75(2):265-7 265-7
DATE:	1993
AUTHOR OF ARTICLE:	Anderson MJ; Townsend DR; Johnston JO; Bohay DR
TITLE OF ARTICLE:	Osteofibrous dysplasia in the newborn. Report of a
ISSN:	0021-9355
OTHER NOS/LETTERS:	Library reports holding volume or year 0014030 8423187
SOURCE:	PubMed
CALL NUMBER:	W1 JO57
REQUESTER INFO:	AB424
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Osteofibrous Dysplasia in the Newborn

Report of a Case*

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Investigation performed at Children's Hospital of Northern California, Oakland

Osteofibrous dysplasia, which has also been called congenital osteitis fibrosa and ossifying fibroma of the long bones, is an unusual disease characterized by eccentric intracortical osteolysis that appears as a single confluent lytic area or as multiple cysts with a honeycomb appearance. It is a benign condition, but it can be mistaken for a malignant lesion.

We examined a newborn who had the striking lytic features in the tibia that are characteristic of this benign condition. We hope that this report will increase awareness of the lesion in the newborn.

Case Report

A newborn infant, who had been delivered by cesarean section (because of a breech presentation) after forty weeks of gestation, had a mass over the anterior border of the left tibia. Five days later, the neonate was referred to Children's Hospital of Northern California.

The newborn had a normal facial appearance, and examination of the musculoskeletal system revealed unremarkable findings, except in the left lower extremity. There was a fusiform mass over the proximal one-third of the anterior aspect of the tibia, beginning distal to the knee. The skin over the mass was normal, and there were no dilated veins overlying the mass. The foot and ankle were normal on inspection and palpation. The ipsilateral knee and hip had a full range of motion. The mass was firm to palpation, and there was no evidence of a bruit on auscultation. The body temperature was 38 degrees Celsius.

Because of the fever, the infant was examined with a complete work-up that included cultures of blood and of specimens obtained with a lumbar puncture, both of which were negative for bacterial growth, and a roentgenogram of the chest, which demonstrated normal findings. On admission to the hospital, the infant had been given ampicillin and gentamicin intravenously, on the assumption that the diagnosis was neonatal sepsis. Administration of the antibiotics was discontinued within forty-eight hours, when the negative results of the cultures had been reported and when the infant was afebrile.

Laboratory investigations included a total and differential white blood-cell count; measurements of the level of hemoglobin, of the sedimentation rate, and of the level of serum alkaline phosphatase; urinalysis; and a serological evaluation for syphilis. The results of all of the studies were within normal limits.

Anteroposterior and lateral roentgenograms of the leg (Figs. 1-A and 1-B) showed an extensive lytic lesion in the middle of the tibial diaphysis with a florid fusiform reactive shell of bone at the periphery. Ossification was observed in the central lytic area. The results of a skeletal survey were otherwise normal so a technetium bone scan was not done. In order to determine the extent and character of the lesion, computed tomography and magnetic resonance imaging of the leg were performed. The computerized axial images (Fig. 2) showed a reactive shell of bone at the periphery of the lytic process in the middle third of the left tibial diaphysis. The T2-weighted magnetic-resonance axial images (Fig. 3) demonstrated high signal intensity in the soft tissues surrounding the thin cortical shell of the tibia, which produced a low signal. High signal intensity was seen throughout the medullary canal.

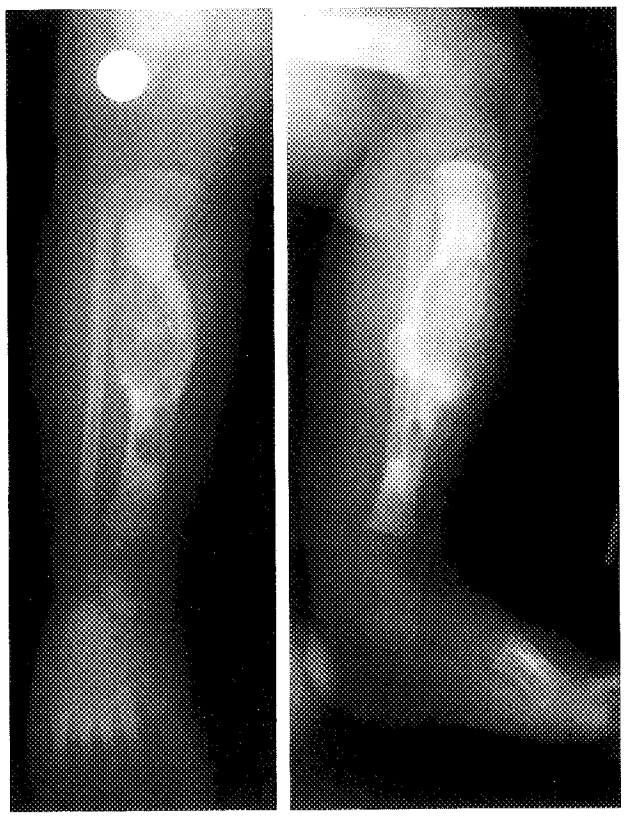

Fig. 1-A Fig. 1-B

Roentgenographic appearance of the leg showing a florid fusiform, expansile lytic destructive lesion in the middle of the tibial diaphysis with a reactive peripheral shell of bone that appears broken anteriorly and laterally. Ossification is visible within the central lytic area.

The differential diagnosis included fibrous dysplasia (the monostotic form), histiocytosis X, congenital syphilis, adamantinoma, and congenital pseudarthrosis (neurofibromatosis). In an effort to determine the diagnosis, an open biopsy, centered over the anterior crest of the tibia, was performed when the neonate was thirteen days old. The removed tissue was whitish-yellow, with a fibrous consistency. Histopathological analysis (Fig. 4) showed benign-appearing woven-bone trabeculae with the characteristic peripheral osteoblastic rimming seen in a benign fibrous-tissue stroma. Microbiological cultures were

*No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article. No funds were received in support of this study.

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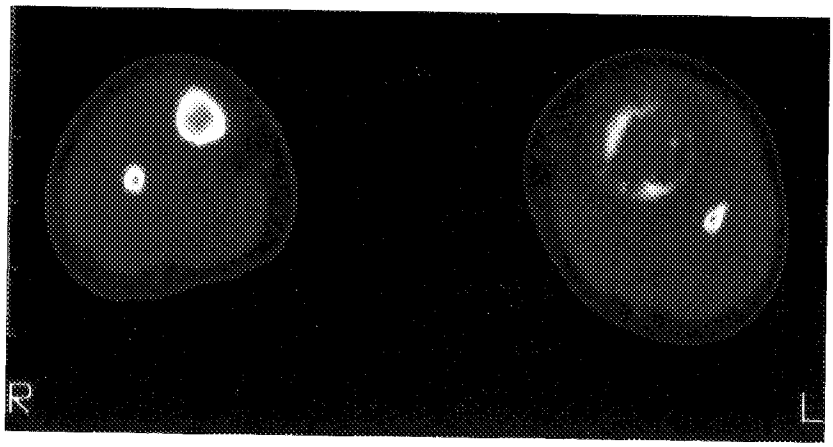

FIG. 2

Computerized axial-tomography images of the right and left legs, demonstrating an expansile lesion with a shell of bone at the periphery, located in the middle of the diaphysis of the left tibia.

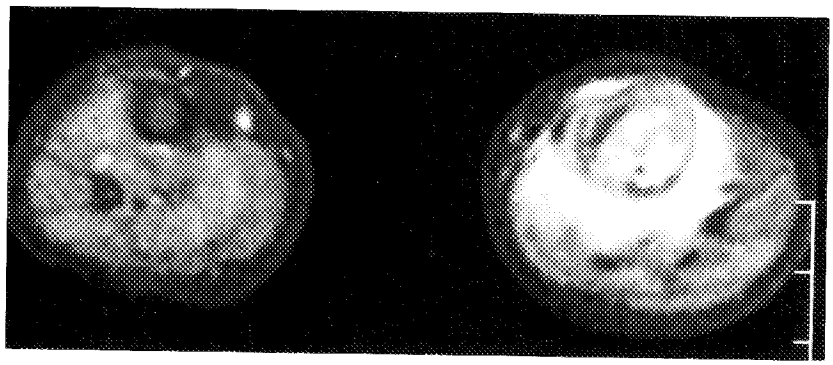

FIG. 3

T2-weighted magnetic-resonance axial images of the right and left legs, showing high signal intensity in the soft tissue surrounding the lesion, with a low signal from the thin shell of bone surrounding the lesion. High signal intensity is also present throughout the medullary canal.

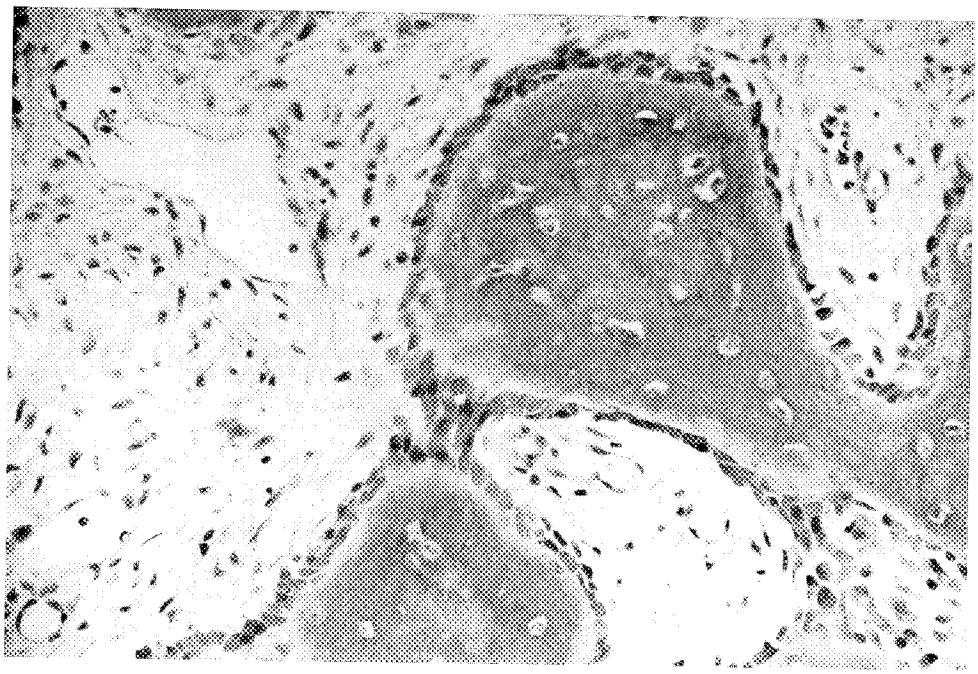

FIG. 4

Biopsy specimen from the anterior tibial neoplasm. There are benign-appearing woven-bone trabeculae with peripheral osteoblastic rimming in a benign fibrous-tissue stroma (hematoxylin and eosin, × 150).

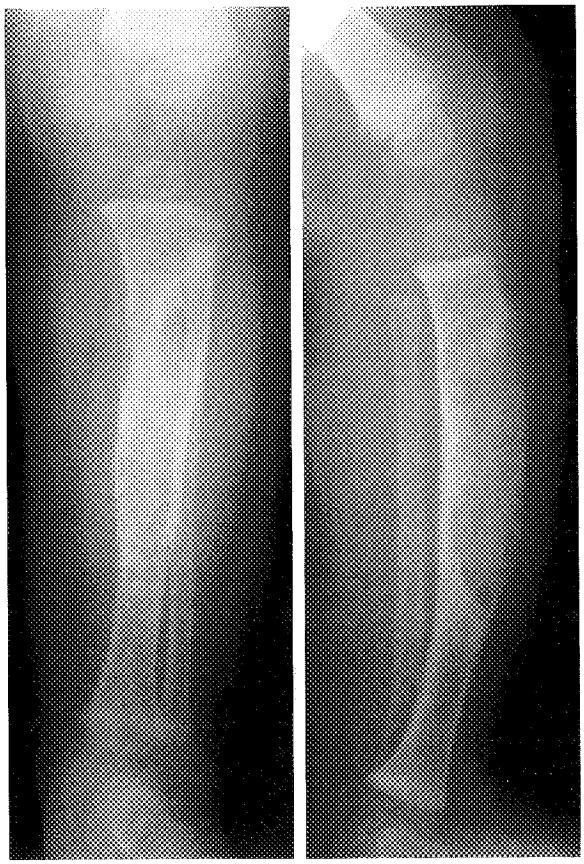

FIG. 5-A FIG. 5-B

Anteroposterior and lateral roentgenograms of the left tibia and fibula, made at the seven-month follow-up, showing remodeling of the bone with filling of the central lytic area and a more normal appearance of the bone. The bowed anterior tibial cortex still has a predominantly smoky appearance, which probably represents persistent dysplastic fibrous tissue

negative for aerobic, anaerobic, fungal, and tuberculous organisms.

After the biopsy, the extremity was immobilized in an above-the-knee cast for eight weeks in order to prevent an iatrogenic fracture. The most recent follow-up roentgenograms, made when the infant was seven months old (Figs. 5-A and 5-B), demonstrated that remodeling had resulted in a more normal anatomical appearance and density of the tibia. Persistent fibrous tissue, evidenced by a smoky appearance of the bone, was still visible in the bowed anterior tibial cortex.

Discussion

Osteofibrous dysplasia was described in 1921 by Franghenheim, who called it congenital osteitis fibrosa. Kempson, in 1966, reported the development of this entity in two patients; he suggested that it be called ossifying fibroma of the long bones because the histological pattern of the lesion was the same as that seen in association with ossifying fibroma of the jaw. The lesion is usually seen only in one tibia, but occasionally the ipsilateral fibula is affected as well. Histologically, the entity resembles those that occur within the mandible. Campanacci[1] described the lesion in 1976 and, in 1981, Campanacci and Laus[2] reported on thirty-five patients. This entity is often referred to as Campanacci disease; usually it is first seen in the first decade of life, and occasionally it is first seen in the first year of life. Although Campanacci and Laus[2] stated that "the symptoms ... are present at or shortly after birth," the youngest patient in their series was three months old.

The differential diagnosis includes fibrous dysplasia, eosinophilic granuloma, congenital syphilis, adamantinoma, and congenital pseudarthrosis. The lesion may be mistaken for fibrous dysplasia because the two entities look similar on roentgenograms; however, there is a great difference between them in terms of the histological findings. With fibrous dysplasia, rimming osteoblasts are not seen around the osseous trabeculae. The histiocytoses, such as eosinophilic granuloma, can be differentiated from osteofibrous dysplasia histologically. Plump bilobed eosinophils characterize eosinophilic granuloma. Congenital syphilis can be distinguished from osteofibrous dysplasia because the former is associated with positive serological markers. Adamantinoma is unusual in the age-group that is affected by osteofibrous dysplasia, and an epithelioid component is usually seen on histological examination. Congenital pseudarthrosis has characteristic clinical signs, and histological examination shows a disorganized hamartomatous cuff at the site of the lesion.

As outlined by Campanacci and Laus[2], treatment of osteofibrous dysplasia in children who are less than fifteen years old should consist of close observation without operative intervention. This recommendation was based on their finding that the tumor recurred locally in sixteen of the twenty-five patients whom they had managed with open curettage and subperiosteal resection. Campbell and Hawk found local recurrence in four patients who had been managed with curettage and autogenous bone-grafting. Those authors recommended non-operative treatment unless the structural integrity of the tibia is threatened and a pathological fracture is imminent.

Despite the rarity of the lesion, it is important to suspect the diagnosis clinically, even when the bone has a markedly lytic appearance, and to confirm the diagnosis with an open biopsy. Once the diagnosis of osteofibrous dysplasia in a newborn has been confirmed, it appears that non-operative management is indicated.

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